# "Mad Cow", Creutzfeldt-Jakob Disease (CJD), and other Transmissible Spongiform Encephalopathies (TSE)

Health Education Facts

**What are TSE?**

Transmissible Spongiform Encephalopathies are a family of prion diseases characterized by spongy degeneration of the brain. This group of similar, but significantly different, diseases infects several different species of animals including humans. (See following table.)

**What is BSE?**

BSE is a slowly progressing degenerative brain disease of cattle. The disease is fatal for cattle within weeks to months from onset of clinical symptoms. BSE was first diagnosed in cattle in the United Kingdom in 1986. One case of BSE in a cow was recently found in the United States.

**What causes BSE?**

BSE is associated with a transmissible agent that affects the brain and spinal cord of cattle. The exact nature of the BSE agent is unknown. One theory is that BSE is caused by an abnormal specific protein, or prion, found in the brain of infected animals. The prion "replicates" causing plaque deposits in the brain. A second theory is that the agent is virus-like. The agent is highly stable, resistant to freezing temperatures, resistant to drying, and cooking at normal temperatures such as those used in pasteurization and sterilization.

| Species | Disease |
|---|---|
| **Cattle** | Bovine Spongiform Encephalopathy (BSE) |
| **Sheep, Goats** | Scrapie |
| **Elk, Deer** | Chronic Wasting Disease |
| **Mink** | TSE in mink |
| **Cats** | Feline Spongiform Encephalopahy |
| **Humans** | Creutzfeldt-Jakob (CJD) (classic and variant), Kuru |

**How is BSE transmitted?**

BSE appears to have originated from scrapie, an endemic spongiform encephalopathy of sheep and goats recognized in Europe since the mid-18th century. Carcasses of livestock rendered into a nutritional supplement and fed to cattle appears to have contributed to the transmission of BSE by ingestion of contaminated feed. Specialized testing have shown that BSE and scrapie are not identical diseases.

**What is classic Creutzfeldt-Jakob disease (CJD)?**

CJD is a slow degenerative human disease of the central nervous system. Prior to the identification of vCJD, classic CJD was known to exist in only three forms. Sporadic cases (85-90% of CJD cases) have an unknown cause and occur throughout the world at a rate of about 1 per 1,000,000 people per year. Iatrogenic cases (<5%) result from the accidental transmission of the causative agent by contaminated surgical equipment or as a result of cornea or dura mater transplants or the administration of human-derived pituitary growth hormones. Familial cases (5-10%) are associated with a gene mutation. The median age of those infected is 65 years, with a duration of illness lasting an average of 4.5 months.

**What is variant Creutzfeldt-Jakob disease (vCJD)?**

vCJD is considered a TSE because of characteristic spongy degeneration of the brain and its ability to be transmitted. vCJD is a new disease that was first described in March 1996. In contrast to classic CJD, vCJD has affected younger patients (median age is 29 years), has a relatively longer duration of illness (median length 14 months), and is strongly linked to exposure, probably through meat products from cattle infected with BSE.

The average incubation period is unknown, but is thought to be as long as 10-20 or more years. Cases of vCJD are very rare, and always fatal. There is no reliable screening test available—diagnosis can only be confirmed during autopsy. The only case of vCJD in the U.S. was found in a person with extensive ties to the United Kingdom, where BSE is found. No cases of vCJD have been linked to eating beef in the U.S.

**What are the clinical signs and symptoms of vCJD?**

Both forms of CJD are marked by the onset of rapidly progressive presenile dementia and progressive motor dysfunction. Variant CJD differs from classic CJD by having more prominent psychiatric symptoms and signs at onset, a longer course and a lack of characteristic EEG findings often found in classic CJD. In addition, there are differences in pathological findings in brain tissue of patients with vCJD.

**Prevention Measures**

To minimize the chances of BSE from entering the United States, severe restrictions were placed on the importation of live ruminants and certain ruminant products from all European countries. This ban also extends to military and government facilities located worldwide. It is also now illegal in the U.S. to feed ruminants feed containing carcasses of other ruminants.

**What measures have been taken to protect the blood supply?**

Several governments, including the U.S., have implemented policies to prevent transmission through blood donations from persons in the incubation phase of vCJD. In January, 2002, the FDA released the "Guidance for Industry—Revised Preventive Measures to Reduce the Possible Risk of Transmission of Creutzfeldt-Jakob Disease (CJD) and Variant Creutzfeldt-Jakob Disease (vCJD) by Blood and Blood Products", that contains recommended donor deferral criteria.

**Is BSE a food borne hazard for travelers to Europe?**

The current risk for infection with BSE agent among travelers to Europe is extremely small, if it exists at all. However, to reduce this possible risk, travelers to Europe and other areas where European beef could be used may wish to avoid beef and beef products altogether, or by selecting solid pieces of beef muscle meat. The prion has not been found in muscle tissue (meat). Milk, dairy products, poultry, and pork are not believe to pose any risk for transmitting BSE agent.

**What about BSE-contaminated surgical instruments?**

Iatrogenic cases of CJD can result from the accidental transmission of the BSE agent by contaminated surgical equipment. The World Health Organization has developed CJD infection control guidelines that recommends destroying instrumentation used in neurosurgery. As this is not practical or cost effective, WHO recommends (when feasible) the use of disposable instruments that come in contact with high infectivity tissue (brain, spinal fluid, and eyes) and low infectivity tissues (cerebrospinal fluid, kidneys, liver, lungs, lymph nodes, spleen, and placenta). These instruments should then be destroyed by incineration.

One of the three most stringent sterilization methods for heat-resistant instruments should be used to reprocess medical instruments that come in contact with high or low infectivity tissues of patients with suspected or confirmed CJD. Patients in which the diagnosis is unclear should be considered as potentially suspected CJD patients until a clear non-CJD diagnosis is established.

**What is the risk of getting vCJD if a vaccine contained bovine derived components?**

There is no evidence to date that vaccines have contributed to the cases of vCJD seen in Europe. Nor is there any evidence that vaccines harbor the BSE agent. The FDA and other Public Health Service agencies believe that the risk of contamination of any U.S. licensed vaccine with BSE agent is remote and theoretical.

**What are the chances of dogs and cats getting a BSE-like disease from eating pet foods with beef by-products?**

Dogs have not been shown to be susceptible to BSE but cats have. The USDA prohibits the importation of pet food from Europe into the U.S.

**Is there any way to confirm if an animal has been infected?**

There is no test to confirm the disease in a live animal. Microscopic examination of brain tissue after death is the primary method used to confirm a diagnosis of BSE.

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